Health Services
LOS ANGELES COUNTY

Expected Practices

Specialty: Rheumatology

Subject: Approach to Inflammatory Myopathies

Date: May 20, 2014

Purpose:

Approach to the diagnosis and initial management of Inflammatory Myopathies

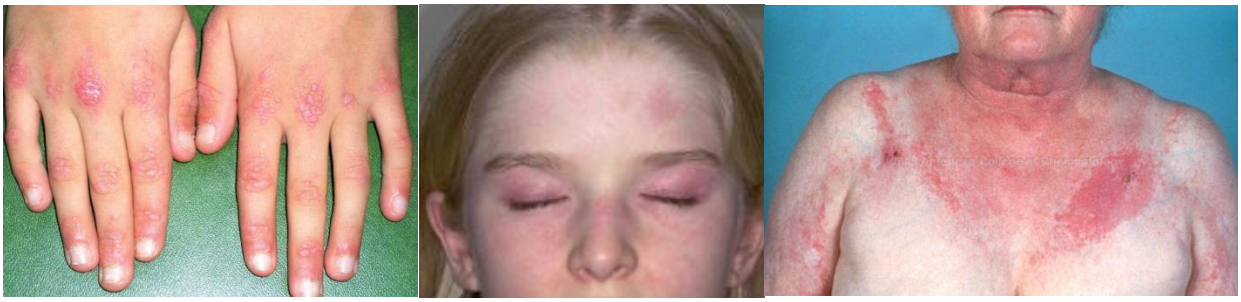
Target Audience: Primary Care Physicians

Expected Practice:

When to think of Inflammatory Myopathies

The inflammatory myopathies include polymyositis, dermatomyositis, inclusion body myositis, and myositis associated with malignancy or other connective tissue disease. These diseases predominantly affect the proximal muscle groups including shoulders, hips, and back. Symptoms of myositis may include trouble rising from a chair or getting out of bed, difficulty climbing stairs or lifting arms, fatigue, and occasionally trouble swallowing or breathing. Weakness, not pain, is usually the predominate symptom. In dermatomyositis, patients may present with a characteristic rash over the MCPs, PIPs, elbows, or knees called "Gottren's papules" (see below). Other dermatologic features include heliotrope rash around the eyes (see below), facial erythema, shawl sign, and V-sign (see below).

This *Expected Practice* was developed by a DHS Specialty-Primary Care Work Group to fulfill the DHS mission to ensure access to high-quality, patient-centered, and cost-effective health care. SPC Work Groups, composed of specialist and primary care provider representatives from across LA County DHS, are guided by 1) real-life practice conditions at our facilities, 2) available clinical evidence, and 3) the principle that we must provide equitable care for the entire population that LA County DHS is responsible for, not just those that appear in front of us. It is recognized that in individual situations a provider's clinical judgment may vary from this *Expected Practice*, but in such cases compelling documentation for the exception should be provided in the medical record.



How to test and risk stratify for Inflammatory Myopathies and when to refer

Start your evaluation with a history focusing on the duration of symptoms. Ask about shortness of breath or difficulty swallowing. Perform a thorough medication and illicit drug/alcohol history and screen the family history for autoimmune diseases or malignancy. On physical exam, test proximal muscle strength including shoulders, hips, and neck flexors. Evaluate the skin for characteristic dermatologic features and examine the lungs for evidence of early interstitial lung disease.

Measurement of serum levels of muscle enzymes, in particular creatine kinase (CK) and aldolase, are very helpful in establishing a diagnosis and following response to therapy. An EMG/NCS in those with elevated muscle enzymes and/or a characteristic history should be performed next and can help establish the diagnosis. At this point in the evaluation, eConsult with rheumatology is recommended. Further testing such as MRI, muscle biopsy and specialized serologic tests should be ordered by the rheumatologist only. Serologic tests including ANA, anti-Jo1, anti-MI-2, and anti-SRP can provide prognostic information but have low sensitivity and are therefore not needed to establish a diagnosis.

eConsult to rheumatology should be made in all patients with a characteristic history, demonstrated weakness on history and exam, and an elevated CK without an obvious precipitant (e.g. statin induced myopathy). For those patients with acute or subacute onset of severe weakness, markedly elevated CK/aldolase, new onset shortness of breath, or difficulty swallowing, prompt referral to the emergency room is indicated for expedited diagnosis and management.

Initial management of newly diagnosed inflammatory myopathy

Discussion with a rheumatologist is indicated prior to initiating therapy. Most inflammatory myopathies are initially managed with high/moderate doses of glucocorticoids followed by immunosuppressive therapies tailored to the severity of the presentation and subset of the disease.